The effect of ventricular septal defect enlargement on the outcome of Rastelli or Rastelli-type repair

Mohammad Ali Navabi, MD,[a] Reza Shabanian, MD,[b] Abdolrazagh Kiani, MD,[b] and Mitra Rahimzadeh, PhD[c]

Objective: Our purpose was to evaluate the effect of ventricular septal defect enlargement on the early and late morbidity and mortality of patients undergoing Rastelli or Rastelli-type operations.

Methods: A total of 49 patients who underwent Rastelli or Rastelli-type operations between 1991 and 2007 were included in a retrospective follow-up study. Patients were divided into 2 groups: group A had ventricular septal defect enlargement, and group B did not have ventricular septal defect enlargement for comparison. Risk factor analysis for early or late death included patient-related and procedure-related variables, with failure, arrhythmia, and atrioventricular block as outcome parameters.

Results: Median age and weight at the time of the operation were 6 years (range, 3 months–22 years) and 17 kg (range, 7–48 kg), respectively. The ventricular septal defect was enlarged in 28 (57%) patients. Ventricular septal defect enlargement showed a significant statistical relation with late ventricular dysfunction, arrhythmia, and residual ventricular septal defect ($P = .023$, $P = .047$, and $P = .01$, respectively, log-rank test). No relation was found between ventricular septal defect enlargement and permanent pacemaker implantation ($P = .73$, log-rank test). Furthermore, enlargement of the ventricular septal defect did not show any significant effect on the rate of early mortality ($P = .69$, Cox regression). Kaplan–Meier estimated survival for patients with ventricular septal defect enlargement was 74% at 5 years and 65% at 10 years. Freedom from late death in the group without ventricular septal defect enlargement was 100% at 5 and 10 years and 83% at 15 years. At a median follow-up of 4 years (range, 6 months–16 years), there were 12 late-onset deaths: 11 in group A (n = 28) and 1 in group B (n = 21). Ventricular septal defect enlargement greatly increased the risk of late death ($P = .009$, Cox regression).

Conclusions: Septal resection in patients undergoing Rastelli or Rastelli-type operations has a substantial effect on late morbidity and is a predictive factor for long-term mortality.

Right ventricle–pulmonary artery conduit interposition, as described by Rastelli for d-transposition of the great arteries with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO), has been used for patients with a variety of heart defects, such as a double-outlet right ventricle (DORV) and pulmonary atresia with VSD. There are several reports regarding the long-term outcome of pulmonary ventricle–pulmonary artery conduit repair. However, these reports mainly focused on survival rate, conduit longevity, and arrhythmia. Little attention was previously paid to the role of VSD enlargement as one of the risk factors for early and late morbidity and mortality. Our aim was to define the influence of VSD enlargement in the early and late outcome of Rastelli and Rastelli-type operations through a retrospective study.

MATERIALS AND METHODS

All patients who underwent Rastelli or Rastelli-type operations between 1991 and 2007 at our institution were included in a retrospective follow-up study. Three patients who were lost to follow-up were excluded. We enrolled patients with 2 different pathologies in which a Rastelli-type conduit for right ventricle–pulmonary artery connection was used: complete transposition of the great vessels and DORV.

A Rastelli-type operation is attributed to procedures using valved conduit for right ventricle–pulmonary artery connection (either a homograft or xenograft) with an intracardiac baffle (Dacron or polytetrafluoroethylene patch) of VSD for connecting the left ventricle to the aorta. All patients underwent operations through a midsternotomy. Standard aortobicaval cardiopulmonary bypass with moderate hypothermia at 25°C and cold antegrade crystalloid cardioplegia were used. We enlarged the VSD in patients with restrictive defects according to the preoperative echocardiographic measurements, which were confirmed intraoperatively with Hegars relative to the aortic valve size by means of direct visualization through a ventriculotomy. The VSD was also enlarged in cases with prominent septal malalignment in which defect baffling would not provide a straight nonobstructive outflow tract (linear flow pattern). It was enlarged by means of incision at the anterosuperior margin of the defect. Including the morphology of Fallot-type DORV in our series might establish the assumption that the subaortic VSD in this group of patients has a significant effect on the technique of intraventricular rerouting and on the late ventricular outflow tract obstruction compared with that seen in patients with transposition of the great arteries morphology. Hence we considered the variable of

From the Department of Pediatric Cardiac Surgery,[a] Imam Hospital, and the Department of Pediatric Cardiology,[b] Children's Medical Center, Tehran University of Medical Sciences, and the Department of Biostatics,[c] Tarbiat Modares University, Tehran, Iran.

Received for publication Sept 26, 2008; revisions received Dec 25, 2008; accepted for publication Feb 16, 2009; available ahead of print May 1, 2009.

Address for reprints: Reza Shabanian, MD, Children's Medical Center, 62 Gharib St, 14194, Tehran, Iran (E-mail: rzshabanian@sina.tums.ac.ir).

J Thorac Cardiovasc Surg 2009;138:390-6
0022-5223/$36.00

doi:10.1016/j.jtcvs.2009.02.034

Abbreviations and Acronyms
AV = atrioventricular
DORV = double-outlet right ventricle
LVOTO = left ventricular outflow tract obstruction
VSD = ventricular septal defect

morphology in the analysis of potential influencing factors for both early and late mortality.

Patients were divided into 2 groups to evaluate the effect of VSD enlargement: group A had VSD enlargement, and group B did not have VSD enlargement for comparison. Early morbidity and mortality were defined as events and death, respectively, within 30 days after the operation. Early reoperation for residual defects was defined as a procedure within 30 days after the operation. Late outcomes for the study were defined as events, reoperation, and death or heart transplantation beyond the first 30 days after the Rastelli operation. Early low cardiac output was defined as the need for 2 postoperative inotropic medications for longer than 30 minutes in the intensive care unit to maintain a systolic blood pressure greater than the lower limit of normal and a cardiac index greater than 2.2 L · min^{-1} · m^{-2} after correction of all electrolyte or blood gas abnormalities and after adjusting the preload to its optimal value. Late ventricular dysfunction (beyond the first 30 days after the Rastelli operation) was defined as low ventricular ejection fraction determined by means of echocardiographic analysis with either the m-mode or Simpson method. The presence of a gradient of 50 mm Hg or greater, as determined by means of echocardiographic analysis, was considered left or right ventricular outflow tract obstruction requiring surgical intervention.

Statistical analysis was performed with SPSS software, version 13.0 (SPSS, Inc, Chicago, Ill). All normally distributed data were expressed as the mean ± standard deviation. The significance of difference between the 2 groups was assessed by using the unpaired Student's *t* test, the χ^2 test, the Fisher's exact test, or the log-rank test, as appropriate. Survival estimates were obtained by using the Kaplan–Meier method. For multivariate analysis, Cox regression was used to establish the variables independently predictive of early and late death. Variables with a *P* value of .1 or less in univariate analyses were entered as candidates into the Cox regression model for determining the significant multivariate predictors. The follow-up status of patients was determined by means of retrospective review of hospital records or telephone interviews. This study was approved by the ethics committee of Tehran University of Medical Sciences. Informed consent was obtained for data extracting from patients' medical files.

RESULTS

A total of 49 patients, including 28 patients with complete transposition of the great vessels and 21 patients with DORV, were enrolled in the study. Twenty-eight (57%) patients had associated pulmonary artery anomalies, including pulmonary atresia, main pulmonary artery stenosis, branch pulmonary artery stenosis, and peripheral pulmonary artery stenosis. The characteristics of patients in groups A and B are depicted in Table 1. The male–to-female ratio in all patients was 33:16, and the age at operation ranged from 12 months to 22 years (mean, 6.8 years; median, 6 years). Nine (18.4%) patients were less than 4 years of age, and weight at the time of the operation ranged from 7 to 48 kg (mean, 19 kg; median, 17 kg). Twelve (24.5%) patients weighed less than 15 kg. Forty (81.6%) patients had a previous palliative procedure, such as modified Blalock–Taussig shunt, septostomy, septectomy, or central shunt. The mean cardiopulmonary bypass time was 179 ± 41 minutes (range, 80–344 minutes), and aortic crossclamp time was 98 ± 22 minutes (range, 47–210 minutes). Forty (82% of patients) homografts and 9 (18% of patients) bovine jugular veins (Contegra; Medtronic, Inc, Minneapolis, Minn) were used in the reconstruction of right ventricular outflow tracts. Patients were divided into 2 groups: group A had VSD enlargement (28 [57%] patients), and group B did not have VSD enlargement (21 [43%] patients). The mean of age and weight at the time of the operation, male–to-female ratio, morphology, palliative procedure, cardiopulmonary bypass time, aortic crossclamp time, and type of conduit were not statistically different between groups A and B (Table 1).

TABLE 1. Comparison of characteristics and perioperative variables in patients with (group A) and without (group B) VSD enlargement

Variable	Group A	Group B	*P* value, χ^2/*t* test
Patients (frequency)	28	21	
Age at the time of operation, y* (mean ± SD)	7.1 ± 4.1	6.3 ± 3.6	.42
Male/female ratio	21:7	12:9	.19
Weight at the time of operation, kg* (mean ± SD)	20 ± 7.8	17 ± 7	.18
Morphology			1
Complete transposition of great arteries	16	12	
Double-outlet right ventricle	12	9	
Pulmonary stenosis/atresia	14	14	.24
MPA, LPA, RPA, or PPA stenosis	11	10	.56
Pulmonary atresia	3	4	.41
Palliative procedure	21	19	.17
Hemoglobin (g/dL)	18.2 ± 4.2	17.8 ± 5.1	.74
Oxygen saturation (%)	70.6 ± 8.1	73.7 ± 7.8	.18
Conduit			
Homograft	21	19	.16
Bovine jugular vein (Contegra)	7	2	.16
Cardiopulmonary bypass time (min)	181 ± 48	176 ± 30	.7
Aortic crossclamp time (min)	99 ± 26	95 ± 18	.5
Cardioplegia (times)	5 ± 1.2	4.7 ± 1.6	.49

VSD, Ventricular septal defect; *SD*, standard deviation; *MPA*, main pulmonary artery; *LPA*, Left pulmonary artery; *RPA*, right pulmonary artery; *PPA*, peripheral pulmonary artery. *Rastelli or Rastelli-type operation.

Early-Onset Morbidity

Nineteen (38%) patients had residual VSD: 15 patients in group A with VSD enlargement and 4 patients in group B without VSD enlargement. Two (4%) patients, 1 in each

TABLE 2. Comparison of early and late morbidity and mortality in patients with (group A) and without (group B) VSD enlargement

Variable	Group A	Group B	*P* value, χ^2/log-rank test
Bleeding	7	2	.27
Early low cardiac output	6	2	.44
Residual VSD	15	4	**.01**
Total reoperation for residual VSD	5	3	.42
Early reoperation	2	1	1
Late reoperation	3	2	.56
Atrioventricular valve damage*	6	1	.09
Early residual LVOTO	0	1	1
Late residual LVOTO	1	6	**.05**
Late residual RVOTO	5	1	**.03**
Complete atrioventricular block	6	5	.81
Transient	3	1	.62
Permanent	3	4	.73
Late ventricular dysfunction	8	1	**.023**
Arrhythmia	7	1	**.047**
Early transient arrhythmia	4	1	.38
Late arrhythmia	3	0	.11
Death	15	1	**.001**
Early	4	0	.125
Late	11	1	**.005**
Infection	1	0	1
Neurologic problem	2	0	.5

VSD, Ventricular septal defect; *LVOTO*, left ventricular outflow tract obstruction; *RVOTO*, right ventricular outflow tract obstruction. *Tricuspid valve.

group, underwent reoperation because of significant residual VSD during the same hospital stay. Early reoperation was also done for postoperative bleeding in 10 (20%) patients, tricuspid valve damage in 7 (10%) patients, and residual LVOTO in 1 (2%) patient. The latter patient was less than 3 years old and in group B. Transient atrioventricular (AV) block occurred in 4 (8%) patients, 3 in group A and 1 in group B; early low cardiac output occurred in 8 (16%) patients, 6 in group A and 2 in group B; transient arrhythmia occurred in 5 (10%) patients, 4 in group A and 1 in group B; convulsion occurred in 2 (4%) patients; and infection occurred in 1 (2%) patient (Table 2). Although residual VSD was significantly related to the enlargement of the defect ($P = .01$, log-rank test), early reoperation necessary for closing a hemodynamically significant residual defect was performed in only 2 patients ($P = 1$, Fisher's exact test). No relation was found between VSD enlargement and transient AV block ($P = .69$, Fisher's exact test). Tricuspid valve damage requiring early reoperation occurred much more in group A (6 vs 1), although it could not be confirmed statistically ($P = .09$, χ^2 test; Table 2). All tricuspid valves were repaired without any valve replacement.

TABLE 3. Predictors for early death

Variable	No.	Event	*P* value, univariate log-rank test	*P* value, multivariate* Cox regression
VSD enlargement	28	4	0.074	.69
Early postoperative bleeding	11	1	0.723	
Atrioventricular valve damage†	7	0	0.399	
Morphology of DORV	21	3	0.04	.71
MPA,LPA, RPA, PPA stenosis, or PA	28	3	0.45	
Age at the time of operation <4 y	9	3	0.003	.97
Body weight at the time of operation <15 kg	12	3	0.015	.98
Aortic saturation <69%	17	1	0.615	
Total cardiopulmonary bypass time >190 min	9	3	0.003	.99
Aortic crossclamp time >100 min	10	3	0.005	.99
Early arrhythmia	5	0	0.486	
Early ventricular dysfunction	8	3	0.001	.30
Transient atrioventricular block	4	1	0.2	

VSD, Ventricular septal defect; *DORV*, double-outlet right ventricle; *MPA*, main pulmonary artery; *LPA*, left pulmonary artery; *RPA*, right pulmonary artery; *PPA*, peripheral pulmonary arteries; *PA*, pulmonary atresia. *Only variables with a univariate P value of .1 or less were entered as candidates into the Cox model. †Tricuspid valve.

Early Mortality

Four (8%) early-onset deaths, all in group A, occurred in the operating room or intensive care unit: 3 patients with DORV and 1 patient with complete transposition of the great vessels. The cause of death was low cardiac output in 2 (50%) patients, bleeding in 1 (25%) patient, and sepsis in 1 (25%) patient. The effect of VSD enlargement on the rate of early mortality could not be shown statistically ($P = .074$, log-rank test). By using multivariate Cox regression analysis, no significant predictor for early death was found (Table 3).

Late-Onset Morbidity

Late ventricular dysfunction (right ventricle > left ventricle) occurred in 9 patients (20% of all survivors): 8 in group A and 1 in group B ($P = .033$, Fisher's exact test; $P = .023$, log-rank test). Three of the 9 patients had late contractility dysfunction with late right ventricular outflow tract obstruction caused by conduit stenosis. Late LVOTO was found in the follow-up of 6 patients in group B (28% of all survivors in group B) and 1 patient in group A (4% of all survivors in group A). All patients were noted to have right bundle branch block. Permanent heart block requiring an implanted pacemaker occurred in 7 patients (15% of all survivors): 3

TABLE 4. Data from patients with late death

Patient no.	Morphology	Palliative operation	Age at Rastelli repair (y)	VSD enlargement	Reoperation early	Reoperation late	Cause of death	Age at death (y)
1	TGA	Yes	6	Yes	AV valve damage*	No	Sudden death (arrhythmia)	17
2	TGA	Yes	13	Yes	No	Conduit replacement	Sudden death (arrhythmia)	28
3	TGA	Yes	4	Yes	No	Conduit replacement	Ventricular dysfunction	15
4	DORV	Yes	5	Yes	No	Conduit replacement	At reoperation for conduit replacement	18
5	TGA	Yes	10	Yes	Bleeding	No	Severe pneumonia	14
6	DORV	Yes	10	Yes	No	No	Sudden death (arrhythmia)	12
7	DORV	Yes	8	Yes	Bleeding	No	Severe pneumonia	12
8	DORV	No	5	Yes	Residual VSD	No	Ventricular dysfunction	5.5
9	TGA	Yes	12	Yes	Residual VSD	No	Ventricular dysfunction	21
10	TGA	Yes	7	Yes	No	No	Ventricular dysfunction	20
11	TGA	Yes	5	No	No	Conduit replacement	At reoperation for conduit replacement	11
12	DORV	Yes	5	Yes	AV valve damage	No	Ventricular dysfunction	6

VSD, Ventricular septal defect; *TGA*, transposition of the great arteries; *AV*, atrioventricular; *DORV*, double-outlet right ventricle. *Tricuspid valve.

patients in group A, who underwent VSD enlargement, and 4 patients in group B without VSD enlargement. No relation was found between VSD enlargement and permanent pacemaker implantation ($P = .73$, log-rank test). Late reoperation was performed in 5 (11%) patients for closing the residual VSD and in 9 (20%) patients for conduit replacement. There was not a significant relation between the enlargement of VSD and late repair for residual VSD ($P = .56$, log-rank test). Freedom from conduit replacement at 5 and 10 years was 80% ± 8.4% and 53% ± 13%, respectively. In 1 (2%) of our patients, redo median sternotomy for homograft replacement resulted in massive hemorrhage caused by the rupture of vascular structures adherent to the posterior sternum.

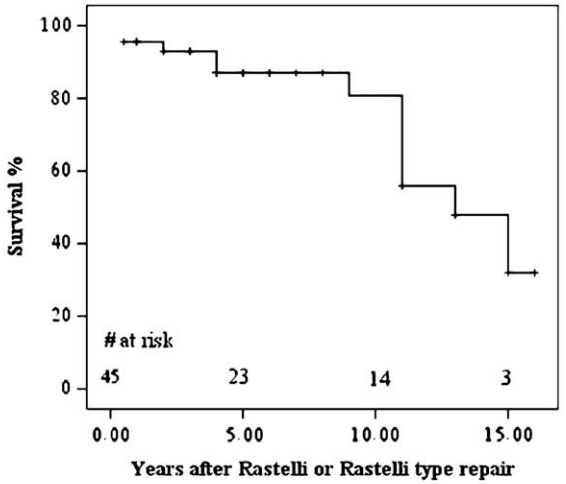
Late Mortality

At a median follow-up of 4 years (range, 6 months–16 years), there were 12 late deaths. No cardiac transplantation was done because of the limited supply and facilities in our country. The causes of late death were sudden death in 3 (25%) patients, ventricular dysfunction in 5 (41%) patients, reoperation for conduit replacement in 2 (17%) patients, and severe pneumonia in 2 (17%) patients. Three late deaths were sudden and unexplained and could have been related to undiagnosed arrhythmias (Table 4). Eleven patients with late death had VSD enlargement at the time of Rastelli repair. Statistically, there was a significant relation between late death and VSD enlargement ($P = .005$, log-rank test). Kaplan–Meier estimated survival for group A at 5 and 10 years was 74% ± 10% and 65% ± 12%, respectively. Freedom from late death in group B was 100% at 10 years and 83% ± 15% at 15 years. The actuarial survival curves for all patients and also each group with and without VSD enlargement are shown in Figures 1 and 2.

Overall, VSD enlargement ($P = .009$, multivariate Cox regression), late ventricular dysfunction ($P = .039$, multivariate Cox regression), late arrhythmia ($P = .034$, multivariate Cox regression), pulmonary artery abnormalities ($P = .035$, multivariate Cox regression), and chronic hypoxemia ($P = .037$, multivariate Cox regression) were significant predictors of late death in our patients undergoing Rastelli repair (Table 5).

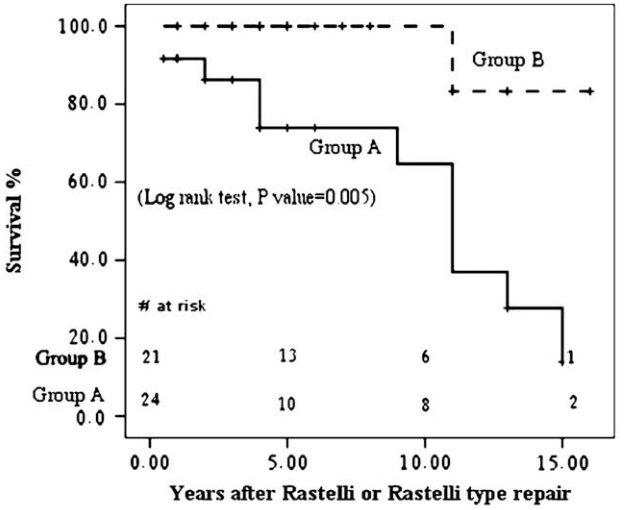
DISCUSSION

The Rastelli operation in its original description consisted of directing the left ventricle to the aorta by suturing a Teflon patch between the VSD and the aortic orifice and of reconstructing the continuity from the right ventricle to the pulmonary artery with a valved conduit.[1] Rastelli suggested diversion of the left ventricular output through the VSD into the aorta. Hence the VSD must be adequate in diameter to allow a completely unobstructed outflow from the left ventricle. The position of the aorta and the tricuspid valve



Time (yrs)	Survival probability	Standard error	Confidence interval (95%)
5	0.862	0.059	0.695-0.943
10	0.801	0.080	0.583-0.912
15	0.310	0.157	0.140-0.572

FIGURE 1. Kaplan–Meier estimated survival for all patients after Rastelli or Rastelli-type repair.



Group A (with VSD enlargement):

Time (yrs)	Survival probability	Standard error	Confidence interval (95%)
5	0.727	0.108	0.453-0.880
10	0.636	0.127	0.342-0.827
15	0.087	0.109	0.001-0.411

Group B (without VSD enlargement):

Time (yrs)	Survival probability	Standard error	Confidence interval (95%)
5	1		
10	1		
15	0.800	0.179	0.204-0.969

FIGURE 2. Actuarial survival curves for patients undergoing Rastelli or Rastelli-type repair with (group A) and without (group B) VSD enlargement. The log-rank test showed a significant relation between late death and VSD enlargement ($P = .005$).

with its subvalvar apparatus must permit placement of the patch to redirect the left ventricular outflow into the aortic root. Enlargement of the defect might be inevitable for this reason.[2] A small restrictive VSD could be enlarged by resecting the conal septum or the anterosuperior margin, creating a straighter left ventricular outflow tract. Moreover, enlargement of the VSD might reduce the risk of postoperative LVOTO. Persistence of LVOTO might be due to restrictive VSD that was not sufficiently enlarged or it might develop later as a result of outflow tract muscular hypertrophy. Another explanation for LVOTO is right ventricular hypertension caused by conduit stenosis. Septal hypertrophy and leftward septal displacement will affect the tunnel shape of the systemic ventricular outflow tract. An abnormal mass/volume ratio of the systemic ventricle caused by removing volume overload after the operation is another risk factor in patients who did not receive the VSD enlargement.[2,3] Residual LVOTO in one of our patients could also be explained by the lower age at the time of repair. Hörer and colleagues,[4] in a retrospective study, reported that age at the time of the Rastelli operation played an important role in the rate of freedom from LVOTO reoperation. Younger age at the time of Rastelli repair implements its effects through limited resection of the septum or limited access to create a straight systemic ventricular outflow tract. The higher rate of freedom from reoperation for LVOTO in our study can be explained by the higher median age at the time of the Rastelli or Rastelli-type operations in our patients (6 vs 3.1 years in the study by Kreutzer and associates[3]).

Resection of the interventricular septum, although advantageous, can be the nidus for deleterious effects that result in early and late morbidity or mortality. The effect of VSD enlargement in the outcome of the Rastelli operation has been different in several reported series.[2,4,5] In the series reported by Moulton and coworkers,[2] VSD was enlarged in 80% of their patients without any effect on early or late mortality. However, similar to our findings, Hörer and colleagues[4] found that enlargement of the VSD in 59% of their patients (57% of our patients) who underwent the Rastelli operation was a predictor for late death or transplantation. The question will arise of whether an alternative procedure, such as a single-ventricle approach or Nikaidoh aortic translocation,[6] would lead to a better outcome and survival.

Residual VSDs are seen more frequently after VSD enlargement.[7] This could be related to the large patch necessary to provide unobstructed flow into the aorta. Anomalous ventricular musculature and tricuspid chordae obscuring the margins of the VSD also increase the risk of a residual defect.[2] Although residual septal defects were much more associated with VSD enlargement in our series, early or late reoperation was only necessary in a few cases with important hemodynamic effects. It has been reported that reoperation for residual VSD has an adverse effect on long-term survival, although this could not be confirmed statistically.[2]

Another deleterious effect of VSD enlargement, which required early reoperation in our patients, was tricuspid valve damage. Tricuspid valve damage caused by VSD enlargement was considered to be all significant new postoperative tricuspid regurgitation that was identified by means of

TABLE 5. Predictors for late death

Variable	No.	Events	*P* value, univariate log-rank test	*P* value, multivariate* Cox regression
VSD enlargement	24	11	0.005	**.009**
Atrioventricular valve damage†	7	5	0.006	.08
Morphology of complete TGA	28	7	0.17	
MPA, LPA, RPA, PPA stenosis, or PA	25	8	0.09	.035
Age at the time of operation >4 y	39	12	0.10	.86
Body weight at time of operation >15k g	36	12	0.038	.98
Aortic saturation <69%	16	8	0.008	.037
Total cardiopulmonary bypass time >190 min	6	4	0.013	.97
Aortic crossclamp time >100 min	7	4	0.024	.98
Arrhythmia (sudden death)	3	3	0.10	.034
Late ventricular dysfunction	9	5	0.01	.039
Permanent atrioventricular block	7	3	0.078	.25

VSD, Ventricular septal defect; *TGA*, transposition of the great arteries; *MPA*, main pulmonary artery; *LPA*, left pulmonary artery; *RPA*, right pulmonary artery; *PPA*, peripheral pulmonary arteries; *PA*, pulmonary atresia. *Only variables with a univariate *P* value of .1 or less were entered as candidates into the Cox model. †Tricuspid valve.

echocardiographic analysis in the absence of right ventricular dysfunction and especially in patients whose restriction across the VSD was secondary to the tricuspid valve tissue. When restriction across an existing VSD is secondary to the AV valve tissue, enlargement of this defect might cause loss of valve function.[8] However, the distortion of the valve mechanism occurs more often because of closure of the VSD rather than its enlargement. Multivariate analysis did not show that early reoperation for tricuspid valve damage had an adverse effect on the long-term survival of our patients.

Both supraventricular and ventricular arrhythmias can be seen with Rastelli-type operations.[9] Our study showed that early transient and late arrhythmias together have a significant relation with VSD enlargement. Furthermore, late arrhythmia was a predictor for late death. Although right ventriculotomy might result in arrhythmogenic potential of the scar, both groups of our patients were similar for this myocardial resection.[10,11] Tateno and coworkers[9] reported older age at the time of operation as a predictive risk factor for supraventricular tachyarrhythmia. However, ventricular tachyarrhythmia or sudden death is also associated with right ventricular hypertension caused by progressive right ventricular outflow tract stenosis. Because late arrhythmia and sudden cardiac arrest had a significant effect on patients' mortality late after Rastelli repair, it is reasonable to monitor the patients for finding any probable dysrhythmia by using Holter monitoring. With the increasing use of implantable cardioverter–defibrillators in younger patients, those children who have survived sudden cardiac death or who have refractory ventricular tachycardia might also benefit from implantable cardioverter–defibrillator implantation.[12]

It is speculated that resection of the anterosuperior margin of the defect carries the potential risk for AV block and scar tissue as a substrate for the arrhythmia. It is unlikely that resection of the anterosuperior margin of the VSD in hearts with ventricular d-looping directly damages the conduction tissue because of its position with respect to the septal defect. However, AV block might occur as a result of trauma to the artery supplying the His bundle.[13] Either transient or permanent AV block occurred in both groups of our study. However, there was no relation between VSD enlargement and either complete or transient AV block.

An important cause of late morbidity in this type of operation would be ventricular failure. Several factors might contribute to the ventricular dysfunction, including septal resection, ventriculotomy, prolonged cardiopulmonary bypass and ischemic time, residual LVOTO, conduit stenosis or regurgitation, and arrhythmia. Consequently, the right ventricle might be affected more than the left ventricle in this type of operation.

The interventricular septum plays an important role in the production of stroke volume. VSD enlargement might disturb the structure/function relationship of the septum and reduce its contribution to the cardiac output of both ventricles.[14] Furthermore, septal resection might affect contractile function through an abnormal interventricular septum with a large prosthetic component (the VSD baffle).[15] It might also damage the septal perforating arteries.[13] Right ventricular hypertension caused by an obstructed conduit will cause contractile dysfunction by impairing left ventricular filling and leftward septal displacement. Cardiac resynchronization therapy with biventricular pacing has been shown to be a beneficial therapy in pediatric and adolescent populations after surgical intervention for congenital heart disease. It has produced acute hemodynamic improvement in ventricular dysfunction postoperatively. In more chronic application, cardiac resynchronization therapy has been shown to improve exercise tolerance and heart failure symptoms and most recently to improve survival as a bridge or alternative to heart transplantation.[16]

Although univariate analysis identified several factors that were associated with early death, including lower age at the time of repair, early ventricular dysfunction, and longer cardiopulmonary bypass and ischemic time, the multivariate Cox model did not establish any independent risk factor for early death.

Furthermore, numerous variables were associated with late death in the univariate analysis (Table 5). However, the multivariate Cox model established 5 important predictors

for late death, including VSD enlargement, chronic hypoxemia, pulmonary artery abnormalities, arrhythmia, and late ventricular dysfunction. Chronic cyanosis caused by the longer palliative treatment with the consequence of myocardial fibrosis at the time of operation might have a negative effect on long-term survival.[3]

Hypoxemia, ventricular outflow tract obstruction, and volume overloading over a prolonged period might explain the lower survival rate in our patients (32% at 15 years vs 52% at 20 years in the study by Kreutzer and associates[3]) with higher median age (6 vs 3.1 years in the study by Kreutzer and associates) at the time of the operation. In the cohort of Hörer and colleagues,[4] there was a trend toward an increased risk for death or transplantation in patients 4 years of age or older at the time of Rastelli repair. Therefore early Rastelli repair in infancy is recommended because of improved late outcome and survival. Kreutzer and associates[3] and Hörer and colleagues[4] showed that longer cardiopulmonary bypass and aortic crossclamp times were predictive factors for late death or transplantation in the Rastelli operation. The cardiopulmonary bypass and ischemic times in our series, although trending higher in the group with VSD enlargement, were not significantly different between the 2 groups. Hence VSD enlargement could not affect the late mortality through prolongation of the cardiac arrest time. Moreover, cardiopulmonary bypass and ischemic times were not identified as predictors for late death in the multivariate analysis in our study. A higher risk for late death in patients who received VSD enlargement might be explained by damaging the septal perforating arteries and the conductive tissues. An interesting comparison will be the one considering similar populations of patients with restrictive VSD and comparing patients with VSD enlargement and biventricular Rastelli repair to those with Fontan palliation without VSD enlargement while considering that a restrictive VSD and consequent subaortic stenosis would be a severe limitation toward Fontan palliation. Another interesting comparison would be the Nikaidoh aortic translocation and biventricular outflow tract reconstruction technique, which could be applied to patients with DORV or transposition of the great arteries with VSD who are not candidates for the arterial switch procedure because of pulmonary (future left ventricular outflow tract) obstruction. However, the outlet septum is also opened into the VSD as a part of this surgical procedure.[6]

Limitations

This study encompasses an intermediate period of follow-up (median, 4 years). Extending the follow-up period almost surely will affect the outcomes. Furthermore, the retrospective design of the follow-up and the small ratio of events per variable concerning the end point of death are other limitation of our study. Patients with malaligned VSD who receive VSD enlargement without ventriculotomy will be a source of data that the exclusive effect of ventricular septal resection could be evaluated by removing the effect of ventriculotomy.

CONCLUSIONS

Septal resection for creating a straight systemic ventricular outflow tract in patients undergoing Rastelli-type operations has a substantial effect on early and late morbidity and is a predictive factor for long-term mortality. Enlargement of the VSD has been associated with a higher prevalence of arrhythmia, late ventricular dysfunction, and residual VSD after Rastelli procedure. Chronic hypoxemia and pulmonary artery abnormalities were other predictors for late death. Hence VSD enlargement is one of the multiple causes of the late mortality in patients undergoing Rastelli operations. However, resection was not associated with an increased requirement for permanent pacemaker implantation. The higher median age of our patients at the time of the Rastelli operation compared with that seen in similar studies could explain the lower survival rate of our patients through prolonged exposure to factors such as hypoxemia and volume overloading.

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